

Fibrous Dysplasia of the Femoral Neck

TREATMENT BY CORTICAL BONE-GRAFTING*

BY WILLIAM F. ENNEKING, M.D.†, AND PETER F. GEAREN, M.D.†, GAINESVILLE, FLORIDA

From the Department of Orthopaedic Surgery, JHM Health Center, University of Florida, Gainesville

ABSTRACT: Fibrous dysplasia of the femoral neck is difficult to treat. In a series of fifteen young patients, ten had a monostotic lesion and five, the polyostotic form of the disease. Twelve patients were first seen with a fatigue fracture. Grafts of cortical bone were used. The objectives of relief of pain, union of the fracture, and prevention of deformity were achieved in all fifteen patients. Two patients required a repeat procedure. None of the patients had important deformity of the femoral neck, and none needed an osteotomy.

The treatment of symptomatic fibrous dysplasia as it affects the femoral neck has received scant attention in the literature. In the polyostotic form of the disease, the well known progressive varus shepherd's crook deformity is associated with shortening, a limp, and occasionally a chronic fatigue fracture with disabling pain. Although monostotic lesions involving the femoral neck seldom produce a severe varus deformity, they are often symptomatic during adolescence or early adulthood. Frequently they are first recognized when a fatigue fracture of the calcar femorale occurs. The purpose of this paper is to present our results with cortical bone-grafting for fifteen patients who had a symptomatic lesion of fibrous dysplasia of the femoral neck. Five of these patients had the polyostotic form of the disease.

Materials and Methods

The study group (Table I) consisted of six male and nine female patients. Their ages at the time of grafting ranged from nine to thirty-two years. Ten patients had monostotic and five had polyostotic fibrous dysplasia. One patient (Case 10) who had polyostotic fibrous dysplasia had acromegaly and precocious puberty and was thought to have a variation of Albright disease. The average duration of follow-up after cortical grafting was six years, with a range from two to fourteen years. The five patients who had the polyostotic form of fibrous dysplasia all had widespread involvement of the skeleton, including both humeri, the digits, and both femora. Of the fifteen patients, all but three

(Cases 2, 4, and 15) had a fatigue fracture when they were first seen. Three patients who had polyostotic disease and one patient who had monostotic disease had sustained more than one pathological fracture of the femoral neck when they were first seen. One patient (Case 3) had first been seen at the age of three years with a fatigue fracture. When she was five years old, the lesion was curetted and packed with cancellous grafts. The lesion recurred, and a second fatigue fracture was evident at the age of eight. A cortical graft was inserted when she was nine years old. Another patient (Case 5) had a fatigue fracture at the age of eleven; it was managed with prolonged immobilization and the use of crutches. The fracture healed, but it recurred within six months after the resumption of activity. Dual fibular allografts were used, as both of the patient's fibulae were dysplastic. Another patient (Case 12) had been seen at the age of seven with multiple lesions, including a lesion in the femoral neck. A biopsy specimen was thought to show an enchondroma. The lesion subsequently was curetted, and the defect was packed with autogenous cancellous bone. She was next seen at the age of twenty-eight years, when she had a symptomatic fatigue fracture and an unresolved lesion of the femoral neck. After the fracture failed to heal despite crutch-assisted walking, the lesion was biopsied again. A diagnosis of fibrous dysplasia was established, and a single cortical graft was inserted.

Surgical Rationale

Our experience between 1960 and 1970 with intralesional curettage of lesions of fibrous dysplasia and autogenous cancellous bone-grafting in various anatomical sites was unsatisfactory in that in the majority of patients, although the grafts were rapidly incorporated, they were subsequently replaced by dysplastic bone (Figs. 1-A through 1-D). The frequency of recurrence of symptoms led us to try an alternative method of treatment. Based on our laboratory experience, we reasoned that autogenous cortical grafts would be much less likely, after incorporation, to be replaced by dysplastic bone than would grafts of autogenous cancellous bone. After creeping substitution has been completed, cancellous grafts are completely replaced by the host's repair bone. In contrast, creeping substitution of autogenous cortical grafts is incomplete; there is replacement only of necrotic osteons, while the interstitial lamellae per-

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† Department of Orthopaedic Surgery, Box J-246, JHM Health Center, University of Florida, Gainesville, Florida 32610.



FIG. 1-A

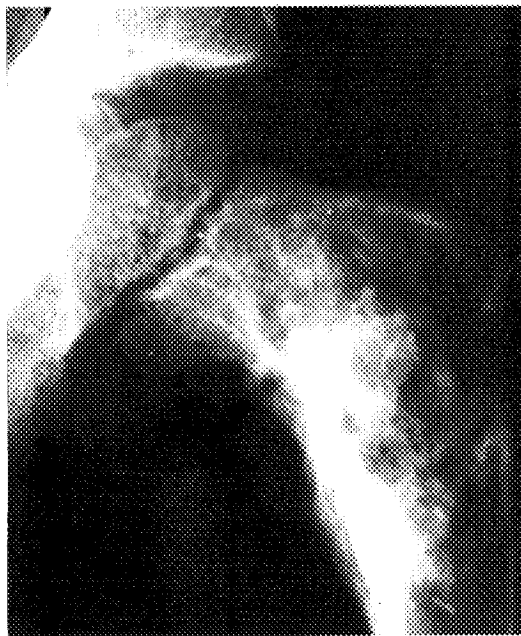


FIG. 1-B

Figs. 1-A through 1-D: Case 11.

Fig. 1-A: This girl had had a monostotic lesion and a fatigue fracture at the age of six years. The lesion was curetted and grafted with cancellous bone. Two years later the lesion persisted, the fracture remained unhealed, the symptoms were still present, and varus deformation had begun. A second curettage and cancellous bone-grafting procedure was done at the age of nine.

Fig. 1-B: The situation remained unchanged, and two years later, at the age of eleven, the fracture had not healed.

sist indefinitely^{2,3,5,6}. Thus, we reasoned that after creeping substitution of a cortical graft in a patient who has a dysplastic lesion, a significant portion of normal matrix — the interstitial lamellae of the cortical graft — should persist and provide structural support.

Clinical Material

Since 1971, we have employed autogenous cortical

grafts in our surgical treatment of patients who have fibrous dysplasia when the goals of treatment have been repair of fatigue fractures and prevention of additional fractures and deformation without aggressive *en bloc* excision. The femoral neck is the most frequent site of symptoms in patients who have fibrous dysplasia of bone, and the symptoms are usually due to a fatigue fracture in the femoral neck. The fracture line itself often is difficult to identify radiograph-

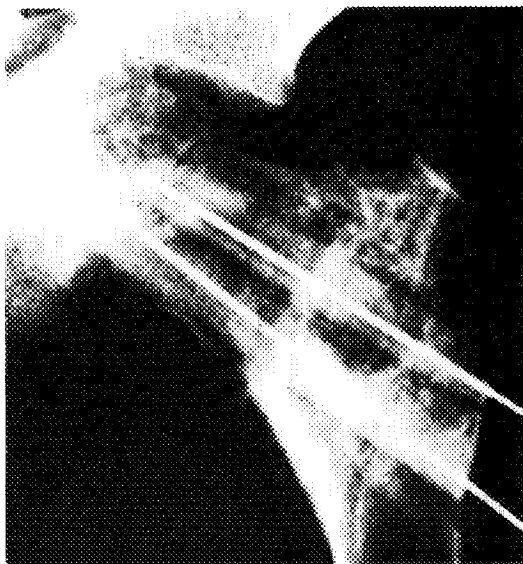


FIG. 1-C

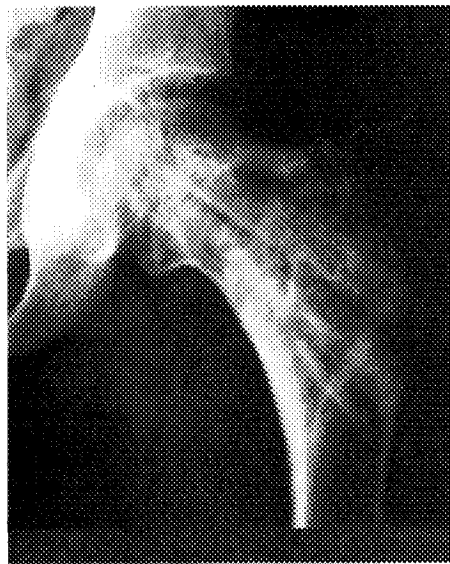


FIG. 1-D

Fig. 1-C: At the age of eleven, dual fibular cortical grafts were inserted. An intraoperative radiograph shows the position of the guide-wires and the grafts.

Fig. 1-D: A radiograph made one year later shows good incorporation of the grafts, healing of the fracture, and persistence of the lesion. The patient remained asymptomatic for the next five years and her activity was unrestricted.

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TABLE I

Case	Age at Grafting (Yrs.)	Sex	Type of Lesion	Prior Surgery*, Age (Yrs.)	Graft	Length of Follow-up (Yrs.)	Fate of the Lesion	Clinical Course
1	20	F	Monostotic†	None	Dual fib.	14	Denser	Asymptomatic; fracture healed
2	21	M	Monostotic	None	Single fib.	8	Same	Asymptomatic
3	9	F	Polyostotic†	Can., 5	Single fib.	6	Denser	Asymptomatic; fracture healed
4	25	F	Monostotic	Can., 16	Single fib.	5	Denser	Asymptomatic
5	15	F	Polyostotic†	None	Dual fib. (allograft)	6	Denser	Asymptomatic; fracture healed
6	11	F	Monostotic†	Can., 8	Dual tib.	8	Larger	Recurrence of pain and fatigue fracture
	19		†	Cort., 11	Dual tib.	5	Smaller	Asymptomatic; fracture healed
7	14	M	Monostotic†	Can., 9	Dual fib.	4	Same	Asymptomatic; fracture healed
8	32	F	Polyostotic†	None	Single fib.	10	Same	Asymptomatic; fracture healed
9	11	M	Monostotic†	None	Single fib.	6	Smaller and denser	Asymptomatic; fracture healed
10	27	M	Polyostotic†	None	Dual fib.	4	Smaller	Asymptomatic; fracture healed
11	11	F	Monostotic†	Can., 6, 9	Dual fib.	5	Same	Asymptomatic; fracture healed
12	29	F	Polyostotic†	Can., 7	Single fib.	2	Denser	Asymptomatic; fracture healed
13	19	M	Monostotic†	None	Dual fib.	3	Denser	Asymptomatic; fracture healed
14	22	M	Monostotic†	None	Dual fib.	2	Smaller	Asymptomatic; fracture healed
15	21	F	Monostotic	None	Dual fib.	1	Same	Recurrent pain
	23			Cort., 21	Iliac cortico-cancellous	5	Denser	Asymptomatic

* Can. = prior cancellous graft and Cort. = prior cortical graft.

† Fatigue fracture at time of cortical bone-grafting.

ically, and its presence may be revealed only after there is formation of callus along the middle portion of the calcar femorale. When a subtle radiolucent fracture line extends across the callus, it gives the appearance of the curved beak of a bird — hence the term parrot beak. *En bloc* excision in a patient who has symptomatic fibrous dysplasia of the femoral neck is seldom indicated, and we have used cortical bone-grafting only in patients who had a symptomatic fatigue fracture or in whom a fracture seemed imminent.

Thirteen patients received their surgical treatment at the University of Florida, and two patients (Cases 6 and 15) were seen only in consultation.

Indications for Operation

The purpose of the graft is to provide a persistent structural span of cortical bone, extending from normal cortex on the lateral aspect of the proximal part of the femur through the dysplastic lesion in the femoral neck into normal cancellous bone in the femoral head. We did not attempt to strengthen lesions that totally involved the proximal part of the femur, where the graft or grafts would be totally encased in dysplastic bone. We restricted the use of this method to patients who had sufficient normal bone in both the femoral head and the lateral aspect of the cortex to anchor the graft

at both ends. Fortunately, in most patients who have fibrous dysplasia of the proximal part of the femur the pattern is one of involvement of the neck and intertrochanteric region, so there is normal bone in the femoral head and the lateral part of the subtrochanteric cortex.

Technique

Using an image intensifier, and with the patient on a standard fracture-table, a straight lateral incision is made to expose the lateral aspect of the femoral shaft and greater trochanter. A guide-wire is introduced at a valgus angle of 135 to 150 degrees that passes through the lesion, so that the proximal tip of the wire is in the normal bone in the femoral head. The wire preferably is situated in the inferior part of the neck, so that the graft, when it replaces the wire, will be juxtaposed to the calcar femorale. A cannulated reamer is inserted over the wire. The outside diameter of the reamer is slightly less than that of the graft. If a biopsy has not been done previously, a small Phemister-type core-punch is used to obtain a specimen. The tunnel is then reamed. The guide-wire can be removed before the graft is inserted or it may be used to guide the graft, in which case it occupies the medullary canal of the fibular graft. Then the graft is inserted. It should fit snugly. If it is in the ideal

position, it extends from the lateral aspect of the cortex, through the lesion, and into the uninvolved femoral head. If a significant amount of subsequent growth from the epiphyseal plate is anticipated, care is taken not to penetrate the plate. Two segments of the fibula (full circumference) are used if the femoral neck will accommodate them. The length of the single or dual fibular graft (or grafts) is calculated from preoperative radiographic measurements. The fibulae are harvested before the tunnel is reamed so that the diameter of the fibula can be measured to determine the appropriate size of the tunnel to ensure a snug fit. No attempt is made to excise any part of the lesion.

The source and number of cortical grafts in the fifteen patients in our series varied. In seven patients, dual autogenous ipsilateral fibular grafts were used, while a single autogenous fibular graft was used in six patients. Dual allogeneic fibular grafts were used in one patient (Case 5) for

whom an autogenous graft was not available due to involvement of both tibiae and fibulae. Dual tibial cortical grafts were used in one patient (Case 6). In two patients (Cases 10 and 13), the lesions involved the proximal third of the femoral shaft as well as the femoral neck, and two fibular grafts were placed in a configuration that was similar to that of the Zickel device (Figs. 2-A and 2-B).

The patient uses crutches for six weeks postoperatively, and partial weight-bearing is allowed. More activity is allowed when the graft is demonstrably united with the lateral aspect of the cortex and with the femoral head.

Results

In all fifteen patients, the continuity and integrity of the graft or grafts were clearly visible on radiographs that were made at the last follow-up visit. Two patients required a second grafting procedure. In one of them (Case 6), two

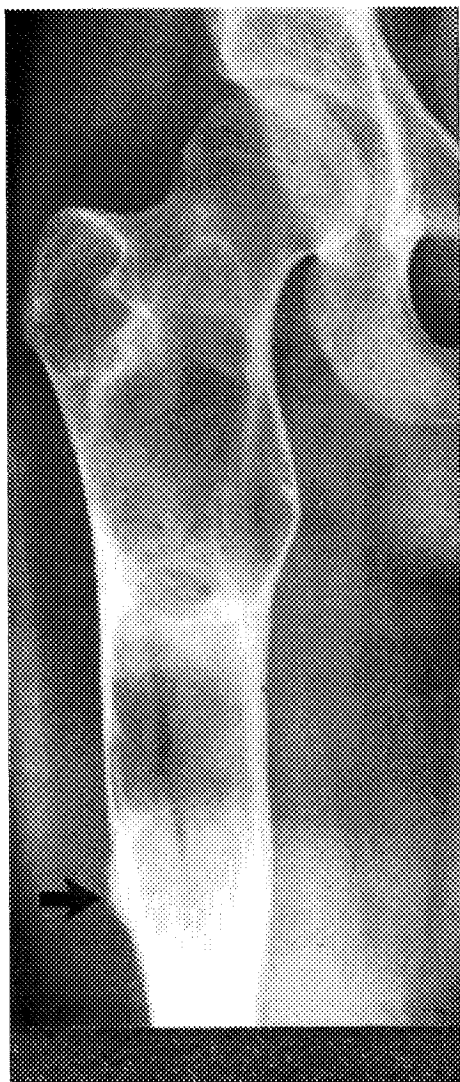


FIG. 2-A

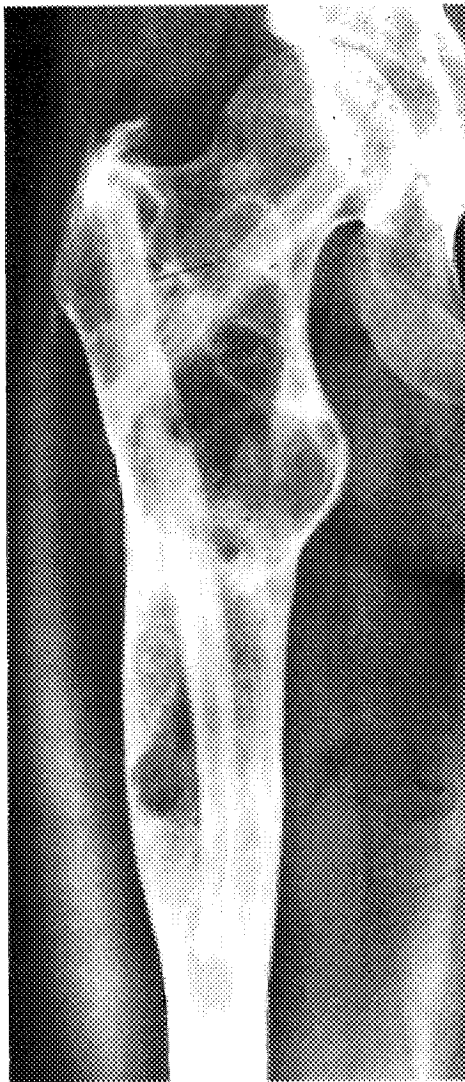


FIG. 2-B

Figs. 2-A and 2-B: Case 13.

Fig. 2-A: A nineteen-year-old man had a monostotic lesion that involved the proximal third of the shaft as well as the femoral neck. A fatigue fracture was present in the distal aspect (arrow). Dual fibular grafts were inserted in a Zickel-like configuration.

Fig. 2-B: Three years later, the fatigue fracture had healed, the symptoms had resolved, the grafts were united and appeared to have hypertrophied, and the lesion was considerably smaller.

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Fig. 3-B

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Fig. 3-C
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Fig. 3-D
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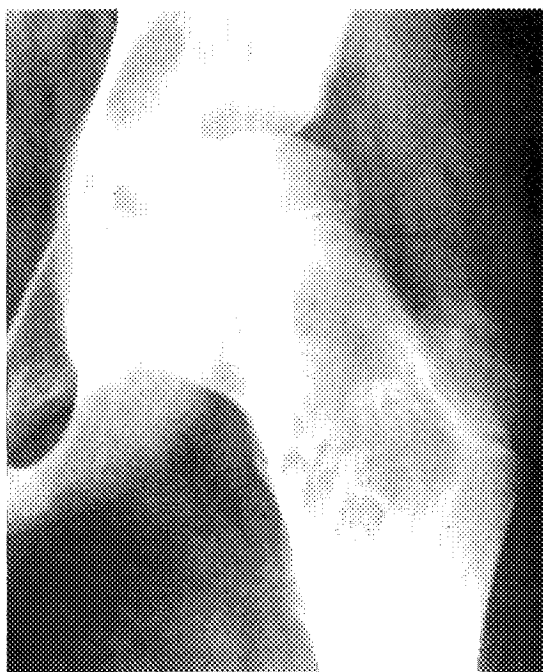


FIG. 3-A



FIG. 3-B

Figs. 3-A through 3-D: Case 6.

Fig. 3-A: An eleven-year-old girl had had a symptomatic fatigue fracture of the femoral neck. Curettage and cancellous bone-grafting had been done three years before.

Fig. 3-B: This time two cortical tibial grafts were inserted, although proximally they did not extend to normal bone.

tibial grafts were initially inserted at the age of eleven (Figs. 3-A and 3-B). The proximal ends of the grafts did not reach normal bone in the femoral head. The patient's symptoms

were relieved when the grafts incorporated, but they slowly resorbed over a period of four years. The lesion gradually enlarged, and eight years later there was a recurrence of

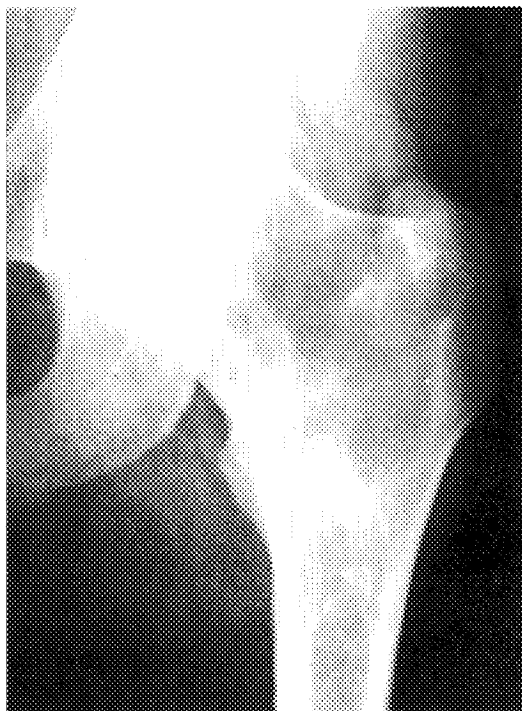


FIG. 3-C



FIG. 3-D

Fig. 3-C: The symptoms resolved, but eight years later, at the age of nineteen, she was again seen with symptoms and a fatigue fracture. At this time the grafts were largely resorbed.

Fig. 3-D: A second grafting with cortical tibial struts was done, this time extending to normal bone. This was followed by union of the fracture and resolution of the symptoms. Five years later, when this radiograph was made, the grafts were intact, the lesion was smaller, and the patient had no restrictions on activity.

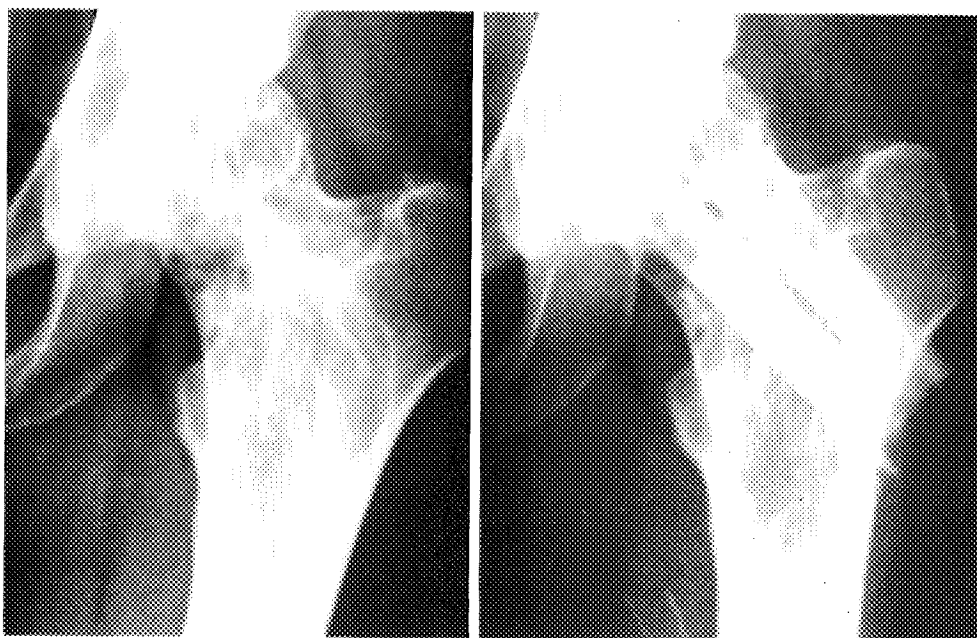


FIG. 4-A

FIG. 4-B

Figs. 4-A, 4-B, and 4-C: Case 1.

Fig. 4-A: A twenty-year-old woman had a fatigue fracture through a monostotic lesion that had never been treated.

Fig. 4-B: Dual fibular grafts were inserted, and one year later there was incorporation of the grafts, union of the fatigue fracture, and relief of the symptoms.

pain with evidence of another fatigue fracture of the calcar (Fig. 3-C). This prompted a second cortical bone-grafting procedure. Five years after this second procedure, the grafts remained intact and the patient was asymptomatic (Fig. 3-D). In the other patient who required a second grafting procedure (Case 15), except for the timing and the source of the grafts the circumstances were similar: initial grafting with dual fibular grafts, resorption within one year, repeat grafting at that time with slabs of corticocancellous iliac bone, and complete consolidation at five years. In the remaining thirteen patients, the grafts have remained intact for two to fourteen years to date (Figs. 4-A, 4-B, and 4-C). In ten patients there was radiographic evidence of diminution in size or increased ossification within the lesion, and the condition of these patients was recorded as improved (Figs. 5-A through 5-D). In only one patient (Case 15) was the lesion completely obliterated. In four patients the size and density of the lesion remained unchanged. In no patient, except for Case 6 after the initial grafting procedure, did the lesion enlarge. In all but one (Case 6) of the patients who were first seen with a fatigue fracture, the fracture remained healed after the initial grafting operation. The fracture line was obliterated by three to six months after grafting. The three patients in whom no fracture could be identified, despite the presence of symptoms associated with a fatigue fracture, became asymptomatic within six months after grafting.

After follow-up ranging from two to fourteen years, there have been no subsequent fatigue fractures in these patients, although radiographically the beak-like projection of callus remained long after obliteration of the radiolucent fracture line. In the patients in whom the lesion diminished

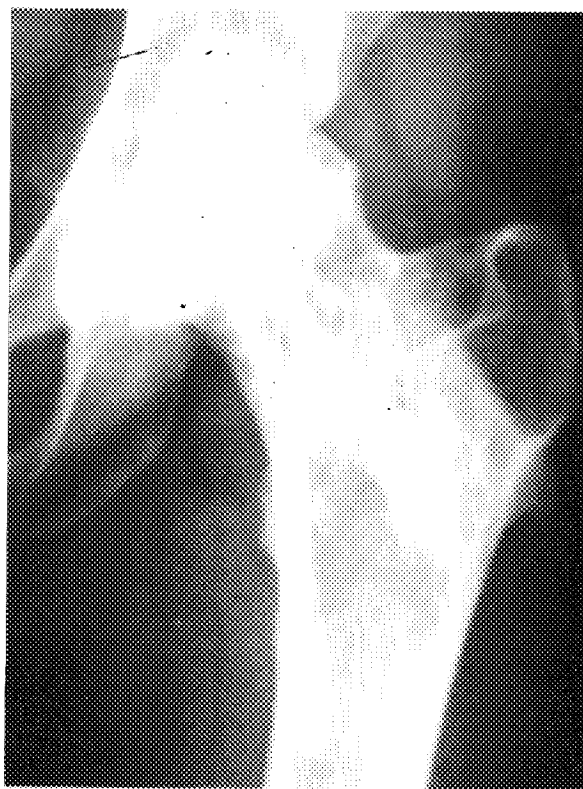


FIG. 4-C

Fourteen years postoperatively the grafts were intact, the calcar had remodeled, and the lesion was denser.

in size, there was enough remodeling of the calcar femorale so that the external remnants of the callus were no longer apparent.

There were no significant complications in these pa-



FIG. 5-A



FIG. 5-B

Figs. 5-A through 5-D: Case 9.

Fig. 5-A: An eleven-year-old boy had a poorly margined lesion and a fatigue fracture.

Fig. 5-B: Insertion of a single fibular graft produced comminution of the dysplastic calcar femorale.



FIG. 5-C

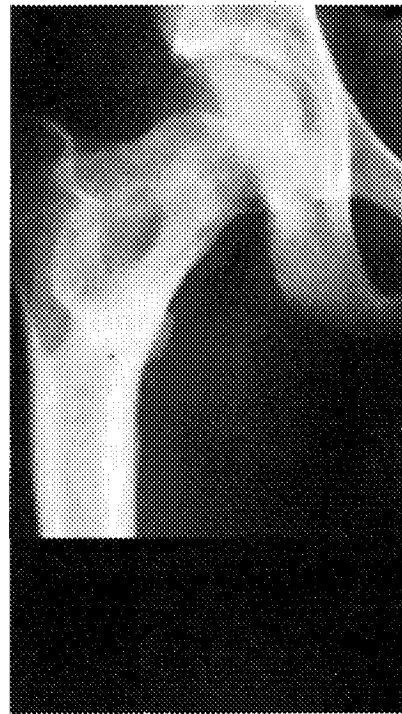


FIG. 5-D

Fig. 5-C: At one year the graft was incorporated, the fracture had healed, the symptoms had resolved, and the lesion was unchanged.

Fig. 5-D: At the age of seventeen, six years after grafting, the grafted calcar had remodeled and the lesion was smaller and denser.

tients, either at the donor site for the graft or at the hip.

Discussion

Pathological fractures and deformities are common in patients who have fibrous dysplasia^{9,11}. The dysplastic bone is biomechanically unsound and fatigue fractures often occur even with normal stresses. Whereas fractures through dys-

plastic bone that occur in the upper extremity or in the vertebrae, causing collapse or scoliosis, or both, may be managed non-operatively, symptomatic involvement of the proximal part of the femur frequently requires surgical intervention to achieve relief of pain and resumption of unrestricted activity. We agree with Jaffe, however, that as to the skeletal lesions: "... the mere fact of their presence

is not in itself an indication for treatment." The age of the patient and the location, size, and biological behavior of the lesion should influence the therapy. The previously reported results of surgical treatment for lesions in the proximal part of the femur have been inconclusive. Harris et al., in 1962, reported four good and five poor results after curettage and cancellous bone-grafting. The Campbell Clinic experience with curettage and cancellous bone-grafting was reviewed by Stewart et al. in 1962. Only six of ten patients who had monostotic involvement of the proximal part of the femur obtained a satisfactory result after an average follow-up of twenty-four months. Thus, of the nineteen reported cases of treatment by cancellous bone-grafting that were analogous to those of our patients, ten were satisfactory and nine failed. When the cases of our six patients who had had prior cancellous bone-grafting are added, the total is twelve satisfactory and thirteen unsatisfactory results in twenty-five patients. It is important to note that the two failures in our series both were in patients in whom the initial cortical grafts did not span the lesion and were not anchored in non-dysplastic bone at both ends.

Other reports in the literature have dealt with patients in whom the indication was correction of a varus deformity

rather than prevention or healing of a fatigue fracture. Funk and Wells, in 1973, reported the cases of four patients who had fibrous dysplasia involving the proximal part of the femur. Two patients who had monostotic disease were managed successfully with a valgus osteotomy and supplementation with a cancellous graft, and two patients who had polyostotic disease required subsequent procedures. Connolly and Breck each reported the successful surgical management of a single patient using a Zickel nail or a long Richards screw-plate without bone grafts. However, the goal in both patients was the correction of an asymptomatic varus deformity. In our patients, the goal was relief of symptoms caused by a fatigue fracture secondary to a dysplastic lesion in a relatively undeformed femoral neck, and grafting is not indicated in such patients when the goal is the correction of an asymptomatic deformity.

Conclusions

The principle of cortical bone-grafting without curettage, excision, or internal fixation in a patient who has a symptomatic lesion of fibrous dysplasia in a relatively undeformed proximal part of the femur appears to be biologically and biomechanically sound.

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